

A Rare Case Report of Renal Replacement Lipomatosis With Co-Existing Xanthogranulomatous Pyelonephritis

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ABSTRACT

Renal replacement lipomatosis is a rare entity. It is the end result of renal atrophy usually secondary to chronic obstructive conditions. Co existent xanthogranulomatous

pyelonephritis is still a rarer entity, which we encountered in our department, which is presented in this report. Clinical importance of this entity is the need to distinguish this condition from fat containing tumors.

Keywords: Calculus, Chronic obstructive, Kidney

CASE REPORT

A 60 year old female came with history of vague right lumbar pain to our surgery department. The patient had been having complaints for more than a year and was initially treated with analgesics, antispasmodics and advised proper bowel habits. This treatment however did not alleviate the patient's symptoms. She came to our hospital after hearing some good reputation hearsay. Clinical examination revealed a vague right lumbar mass. Lab investigation revealed normal renal, hepatic parameters, increased blood lymphocyte count and increased urine pus cells. She was referred to the radiology department and subjected to radiological investigations starting from abdominal X-ray.

X-ray showed absence of normal right renal contour and a large dense cloud like calculus in the right renal region. Another radio opaque calculus was noted in the right lower ureteric line [Table/Fig-1].

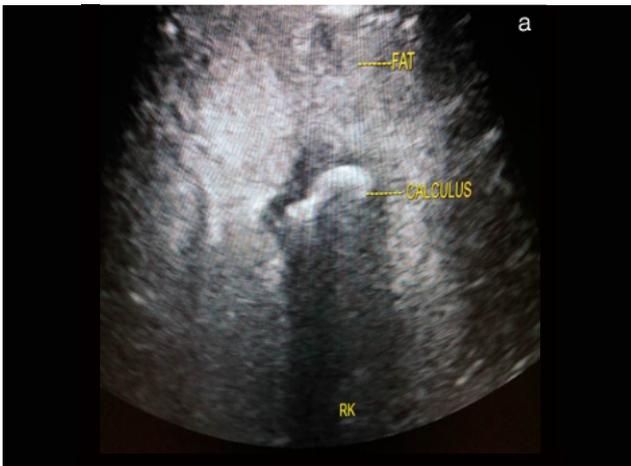
Then the patient was subjected to abdominal USG (ultrasound) [Table/Fig-2A] which revealed absence of right renal architecture. Right renal region showed an ill-defined uniformly hyperechoic mass with central arcuate shaped posterior shadowing. Attempts were futile in locating any ectopic location or atrophy of the right kidney. Left kidney was hypertrophied (11.9 x 5.5 cm) [Table/Fig 2B].

Sensing some rare condition we immediately convinced the patient to undergo CT abdomen and got her consent for undergoing further investigation and be a part of our study. Then the patient was subjected to CECT whole abdomen.

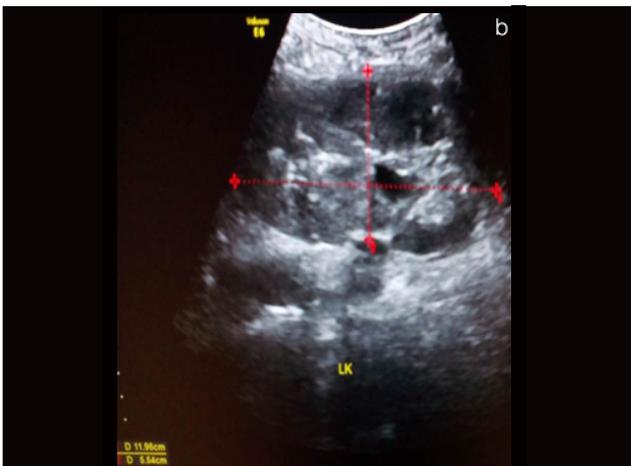


[Table/Fig-1]: X-ray erect abdomen showing absence of normal right renal contour and a large dense cloud like calculus in the right renal region

The topogram confirmed the X-ray findings and the ureteric calculus was more clearly seen [Table/Fig-3a]. The non-enhanced scans [Table/Fig-3b,3c] showed an ill-defined adipose mass filling up the right renal fossa merging with the fat of the retroperitoneal compartments. Radiating soft tissue strand were noted randomly within the fatty mass. A small pocket of air pocket containing collection was also noted within the right renal region. A large dense calculus was noted within the right mid ureteric lumen, with upstream hydro ureter.



[Table/Fig-2a]: B Scale USG image showing an ill-defined hyper-echoic mass with central arcuate shaped posterior shadowing in the right renal region



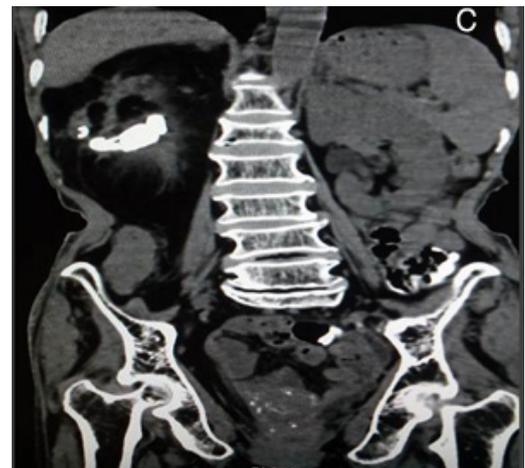
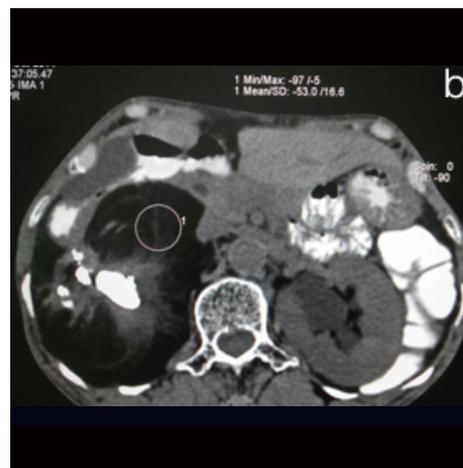
[Table/Fig-2b]: B Scale USG image showing hypertrophied left kidney



[Table/Fig-3d]: CECT axial image showing a small peripherally enhancing collection with internal air focus

On post contrast scans the soft tissue strands (the remnant renal tissue) did not show any perceivable enhancement [Table/Fig-3d]. There was no contrast excretion in the right renoureteric system, even in the 24 hours delayed scan. Small peripherally enhancing collection was noted with internal air focus suggesting coexisting chronic infection which was also supported by increased pus cells in urine analysis. Putting all the findings together, we went for a USG guided cut biopsy which confirmed mature adipose tissue with sparse atrophied interspersed renal tissue on histopathological examination (HPE).

In this case as we have a causative factor in the form of chronic ureteric obstruction and radiologically no detectable enhancing soft tissue and non-malignant HPE result, we came to the diagnosis of Renal Replacement Lipomatosis (RRL). The peripherally enhancing collection within the right renal region which favours coexistent Xanthogranulomatous Pyelonephritis (XPN).



[Table/Fig-3a]: CT Topogram showing enlarged right renal shadow with cloud like calcification. Another calculus seen in the right lower ureteric line **[Table/Fig-3b,c]:** Axial and coronal NECT images showing an ill-defined adipose mass filling up the right renal fossa merging with the fat of the retroperitoneal compartments with internal soft tissue strands

The patient did not give consent to undergo any form of surgical procedures. Due to the compensatory overload of the left kidney, it will be prudent to keep the renal parameters within normal limits as possible. After discussing with surgeons, physicians and urologists, we decided to follow up the patient. She is on anti hypertensives and other reno protective agents.

DISCUSSION

Replacement lipomatosis is an advanced form of renal sinus lipomatosis which occurs in many obese elderly people. The word replacement is used since the renal tissue is entirely replaced with fatty tissue. Along with fat, fibrous tissue also gets laid down resulting in a non-functioning kidney. Interestingly many cases demonstrated in literature was secondary to distal obstruction and our case is no exception. But the rare feature in our case is the co-existing pyelonephritis as evidenced by peripherally enhancing collection and increased urine pus cells. The co-existing diagnostic consideration in the presence of long standing inflammation and calculus obstruction (as in this case) is xanthogranulomatous pyelonephritis [1, 2].

The main differential diagnosis for this condition is fat containing neoplasms. It is very difficult to rule out underlying neoplasm in this case. However the limited specimen got through the cut biopsy revealed mature fat cells. The differential diagnosis includes: angiomyolipoma, lipoma, and liposarcoma [3].

Radiologically it is challenging to differentiate RRL from fat containing neoplasms. However we can take a clue from the fact, in RRL the fat is more concentrated in the original location of the sinus [4]. Renal replacement lipomatosis is a chronic condition which has no curative treatment. Some authors describe radical nephrectomy and post-surgical close follow up to rule out remote possibility of malignant transformation [5].

CONCLUSION

Renal replacement lipomatosis is a rare condition and with co existent xanthogranulomatous pyelonephritis is still a rare entity. The patient details are recorded and have advised the patient for regular follow up every six months. The diagnosis and management of this entity is based on a focussed radiological approach with good support from the clinical side.

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